Information

Congenital Heart Disease In the First Year of Life

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SURGICAL ADVANCES in the past two decades in the management of patients with congenital heart defects have been truly spectacular. These results have been achieved for the most part in older children and adults. It is somewhat sobering to note, however, that 95 percent of deaths due to these defects occur in the first year of life, most in the first six months. Twenty percent of patients with congenital cardiac defects develop congestive heart failure; 90 percent of these patients are less than one year of age. The present thrust must be directed toward the treatment of the young infant. To ensure success, an aggressive approach is necessary in which the physician supplying "first-line" care (generalist or pediatrician) becomes a participant in the team which includes the pediatric cardiologist and cardiovascular surgeon. Pediatric cardiologists and cardiovascular surgeons are, for the most part, aware of their role. The "first-line" physician needs to recognize his role as an active participant so that infants in jeopardy will be recognized and treated early or the total endeavor is doomed to failure.

A small number of anomalies are responsible for congestive heart failure or death in most infants. Palliative or corrective surgery in infancy is available for infants with anomalies listed in Table 1, while there is no surgical relief for those with anomalies listed in Table 2, as well as in some of the more complicated lesions seen less frequently.

It is not essential for the generalist to make an exact anatomical diagnosis; this is required of the pediatric cardiologist. Identification of the infant in early heart failure and those with a predisposition to hypoxic "spells" or sudden death is the generalist's chief role. This will enable him to identify the infant requiring emergency or early referral to a center equipped and staffed to perform the necessary diagnostic studies with a minimum of risk to the infant.

All infants known to have a congenital cardiac anomaly should be followed closely during the first six months, their period of greatest risk. Most do not require referral to a large center. The thriving asymptomatic infant should have an electrocardiogram and x-ray of the chest performed at about one month of age. If these are normal, subsequent referral may be indicated on an elective basis.

Emergency referral to a center is indicated for infants who show signs of congestive heart failure or in whom the electrocardiogram reveals a "strain" pattern of either left or right ventricle. In the cyanotic infant, emergency referral is indicated in those with a history of hypoxic "spells," respiratory distress, evidence of pulmonary plethora in the chest film or right ventricular "strain" in the electrocardiogram. Procrastination results in decreasing myocardial function resulting from hypoxia and acidemia.

Congestive heart failure in the infant is usually insidious in onset. Decreased cardiac output results in fatigue usually manifested by feeding difficulty. The infant sucks eagerly, but fatigues after

TABLE 1.—*Anomalies Amenable to Surgical Correction*

Acyanotic	*Cyanotic*
Vent. Septal Defect	Tetralogy Fallot
Atr. Septal Defect	Transposition Complex
Patent Duct. Art.	Tricuspid Atresia
Coarctation Aorta	Pulmonary Atresia
Pulmonic Stenosis	Total Anom. Pulm. Veins
Aortic Stenosis	

TABLE 2.—*No Available Surgical Relief*

Hypoplastic left heart syndrome
Endocardial fibroelastosis

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taking a few ounces of formula. He soon becomes hungry, takes a few more ounces and falls off to sleep. A vicious cycle develops, easily confused with intolerance to formula, "colic" and "failure to thrive." Grey, mottled cold extremities indicative of vasoconstriction are a homeostatis response to an inadequate cardiac output.

Tachypnea is usually the earliest manifestation of heart failure in the infant. Pulmonary venous congestion secondary to left heart failure is manifested by tachypnea. Respiratory rates over 50 a minute are abnormal in the quietly resting infant. Those in failure commonly exhibit respiratory rates of 60-100 a minute, and at times, 120-150 a minute. In addition to tachypnea, one may note intercostal and sternal retraction and eventually bronchial congestion with cough and bronchospasm. Rales, in the absence of infection, are only seen in late stages of heart failure. Tachypnea is accompanied by tachycardia and often a gallop rhythm heard best about 0.10 seconds after the second sound, often resembling a third heart sound. Pulmonary venous congestion may be identified on the x-ray film by hilar shadows, at times extending to the periphery of the lung.

Systemic venous congestion secondary to right heart failure is manifested by hepatomegaly, the liver edge extending more than 4 cm below the costal margin. Tenderness is uncommon. Pitting edema and neck vein distention are not usually apparent.

Heart failure of sudden onset is uncommon in infants. It may result from an acute myocarditis, dysrhythmias or myocardial infarction.

"Spells" are seen most often in infants with tetralogy of Fallot. About 10 percent end in fatalities and an equal number are complicated by cerebrovascular accidents. "Spells" are characterized by sudden onset of dyspnea, irritability and a marked increase in degree of cyanosis. The infant becomes limp, may convulse and/or lose consciousness. Spells are associated with temporary disappearance of a pre-existing murmur, decreased arterial oxygen saturation and ST depression with increased p wave voltage in the electrocardiogram. Spells may be seen in very early infancy, becoming most common around nine months of age and decreasing in frequency after eighteen months. They often occur without warning but may be precipitated by crying, fever or hot weather. "Spells" should be treated by placing the infant in the knee-chest position, administering oxygen and morphine (0.2mg/Kg). Urgent referral for diagnosis and surgical relief is indicated.

The possibility of sudden death due to critical aortic or pulmonic stenosis should be considered in any infant with electrocardiographic evidence of left or right heart "strain" patterns. It behooves the generalist to recognize these patterns as his minimum requirement in pediatric electrocardiography.

Several types of cyanosis may be seen in infants. Peripheral cyanosis due to vasoconstriction is associated with cold, mottled extremities. Harlequin cyanosis has no clinical significance, being common in normal infants during the first week of life, and may be seen as late as 28 days. It is confined to one half of the body with strict midline demarcation. More commonly, it is manifested by pallor or redness but semicyanosis may be the presenting feature. Cyanosis due to pulmonary disturbances or right to left intracardiac shunting may be difficult to differentiate on clinical grounds if accompanied by respiratory distress. In the absence of respiratory distress the cause is most apt to be a right to left shunt. We believe that an anatomical diagnosis should be made in all cyanotic infants, but not necessarily on an emergency basis.